

# 第34回神経放射線ワークショップ

## Case presentation



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# Case

【Patient】

18-month-old girl

【CC】

Seizure

【History】

She had common cold-like symptoms for 1 week. She was transferred to the clinic with **generalized tonic - clonic seizure**. CT imaging showed brain tumors, bringing her to our hospital.

【Physical exam】

HR: 170/min, BP: 100/53mmHg, BT: 38°C

Recognize her mother as same as usual

Keeping sitting position

No remarkable abnormal findings in Chest or Abdomen

【PMH】

NA

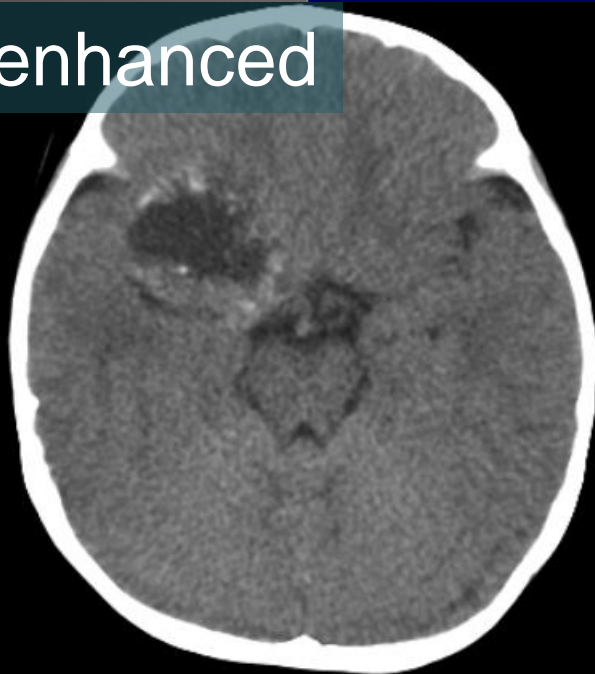
# Blood test

WBC	<u>11730</u>	/μl	TP	6.5	g/dl	CEA	2.2	ng/ml
RBC	435×10 <sup>4</sup>	/μl	ALB	4.3	g/dl	AFP	2.7	ng/mL
Hb	12.0	g/dl	AST	<u>44</u>	IU/l	CA19-9	<u>125.7</u>	U/ml
Hct	35.2	%	ALT	18	IU/l	CA125	33.5	U/ml
Plt	27.8	万/μl	LD	<u>343</u>	IU/l	NSE	<u>29.9</u>	ng/ml
Na	136	mEq/l	T-Bil	0.8	mg/dl	【ABG】		
K	4.4	mEq/l	ALP	<u>714</u>	IU/l	pH	7.422	
Cl	104	mEq/l	Γ-GTP	11	IU/l	PO2	29	mmHg
			Cr	0.20	mg/dl	Lac	13	mg/dl
			BUN	7.3	mg/dl			
			ChE	358	mg/dl			
			NH3	46	μg/dl			
			glu	92	mg/dl			

# Initial imaging

Unenhanced and Contrast enhanced  
CT and MRI

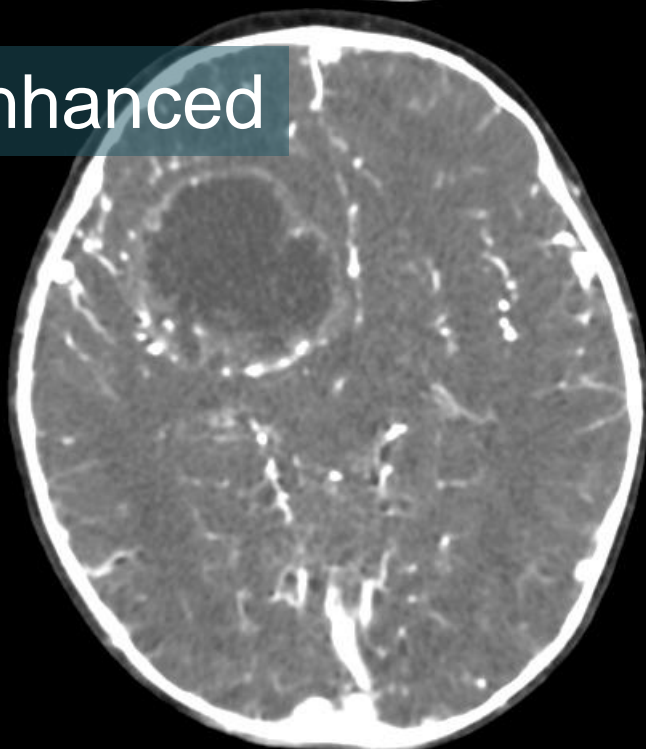
Unenhanced



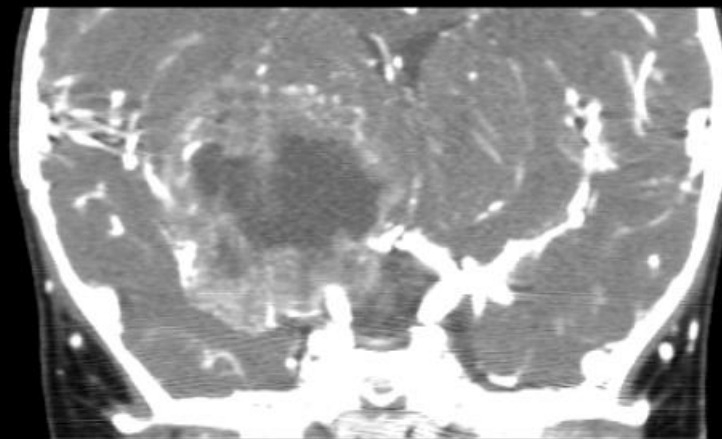
Unenhanced



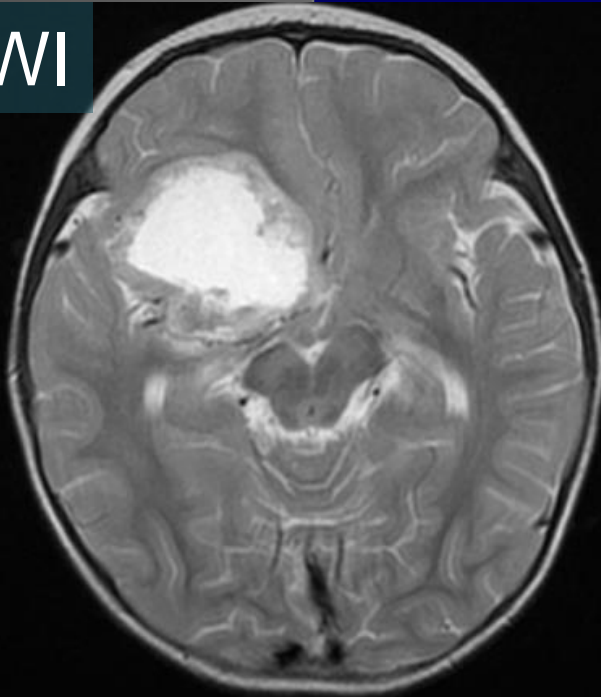
Enhanced



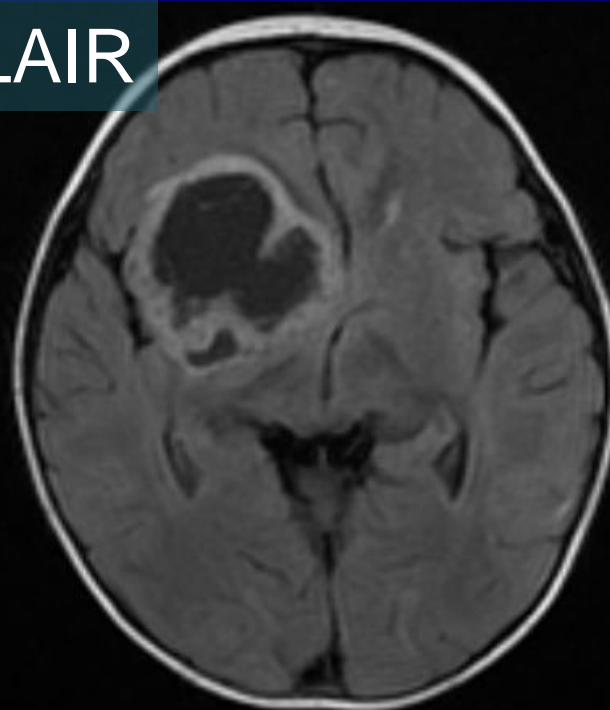
Coronal (MPR)



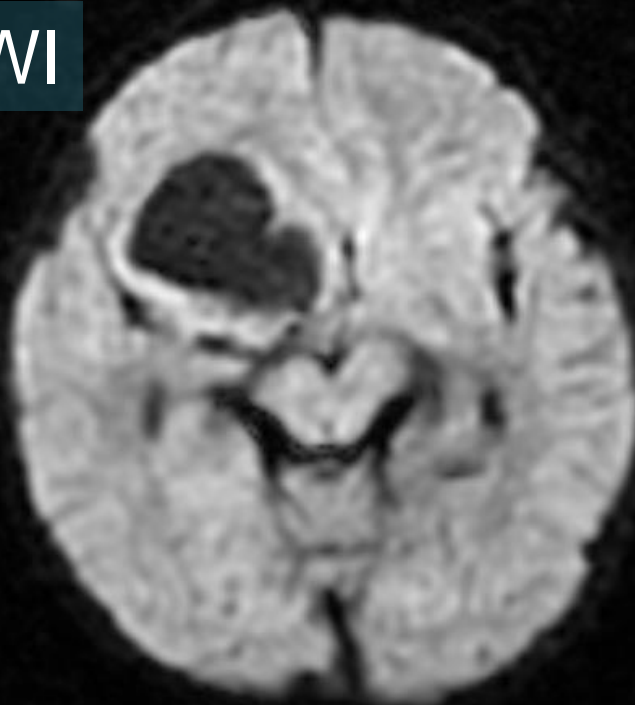
T2WI



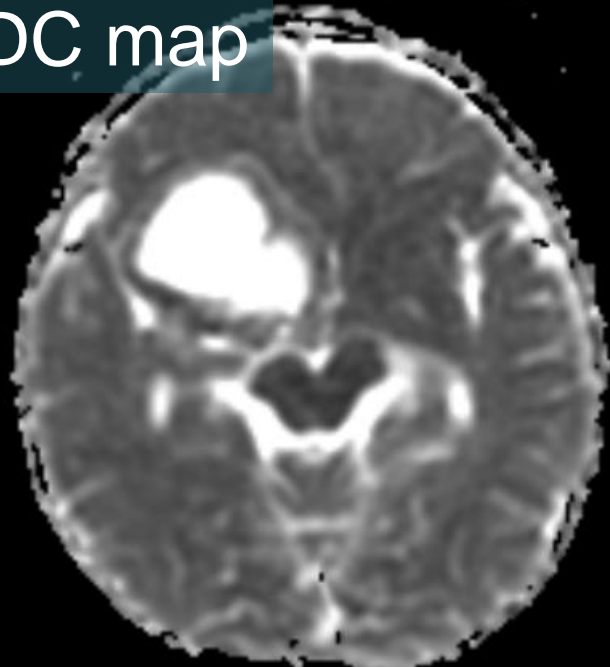
FLAIR



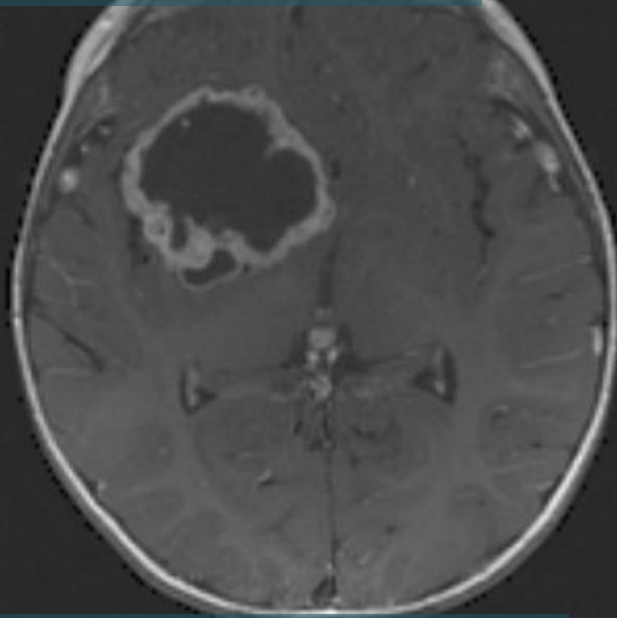
DWI



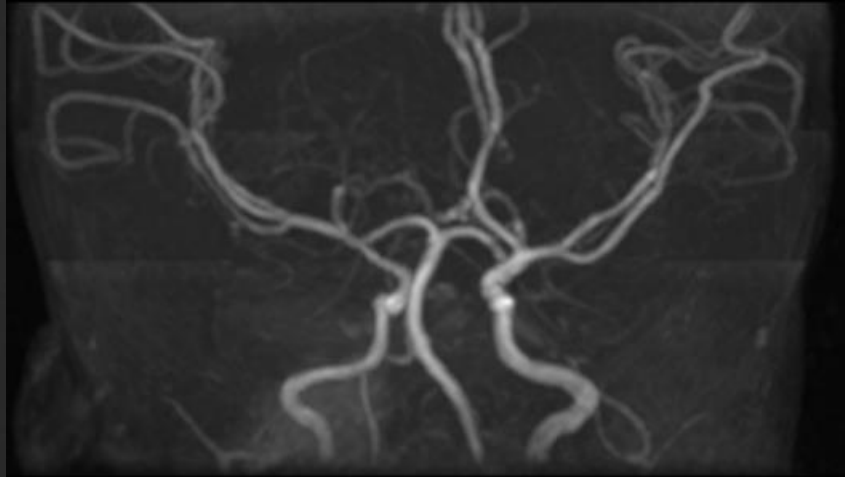
ADC map



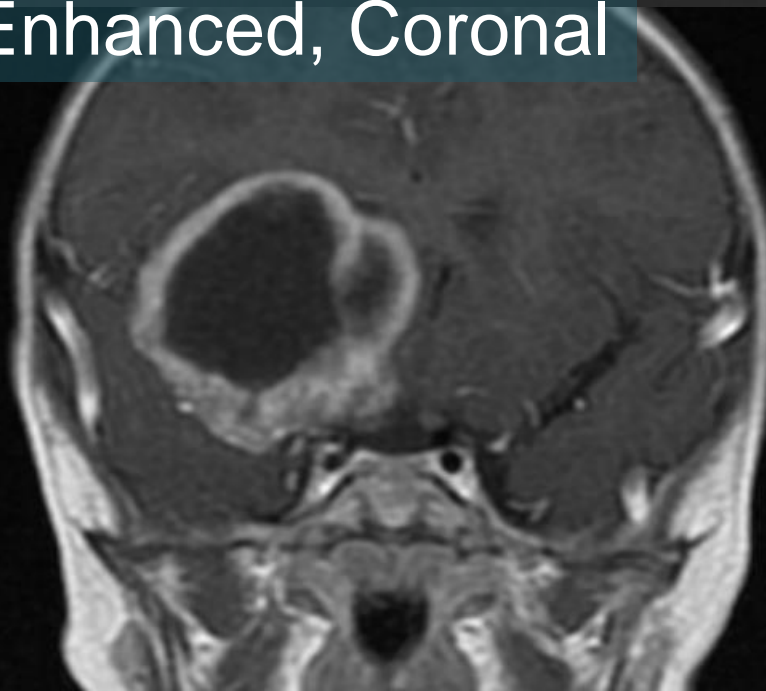
Enhanced T1WI



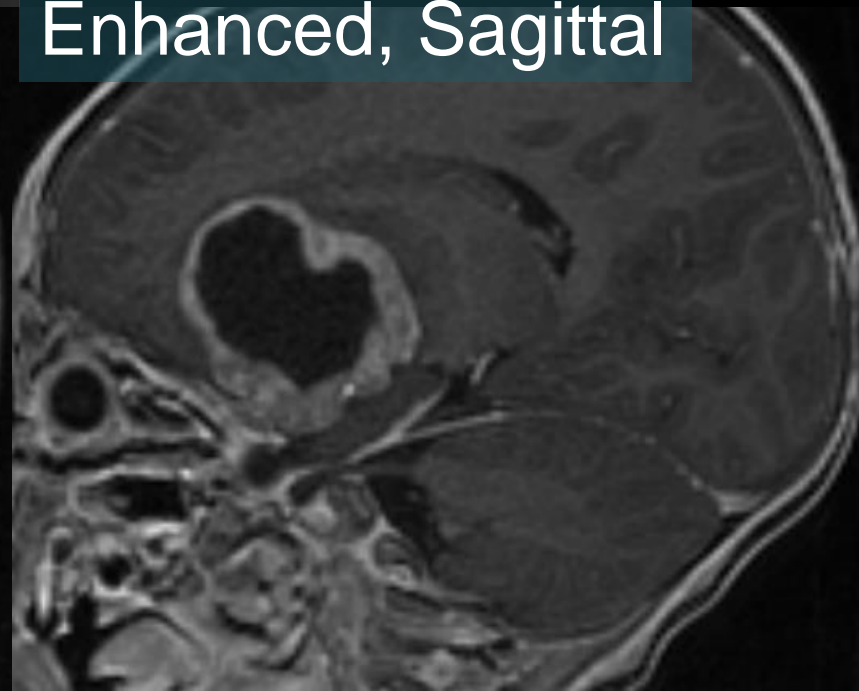
MRA



Enhanced, Coronal

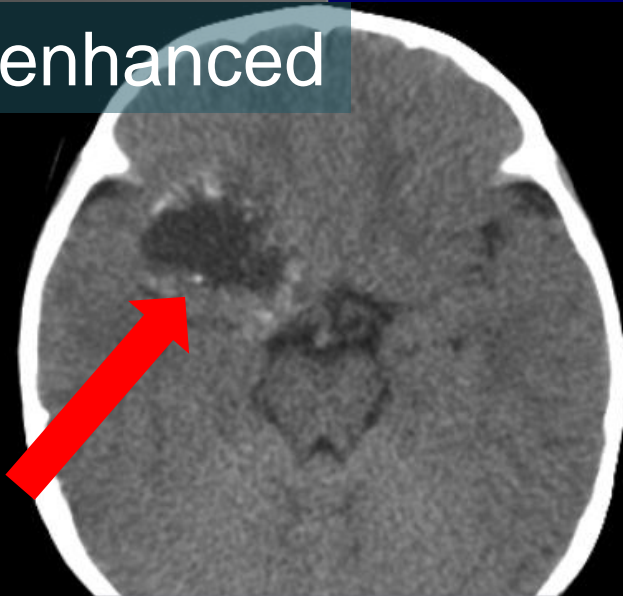


Enhanced, Sagittal



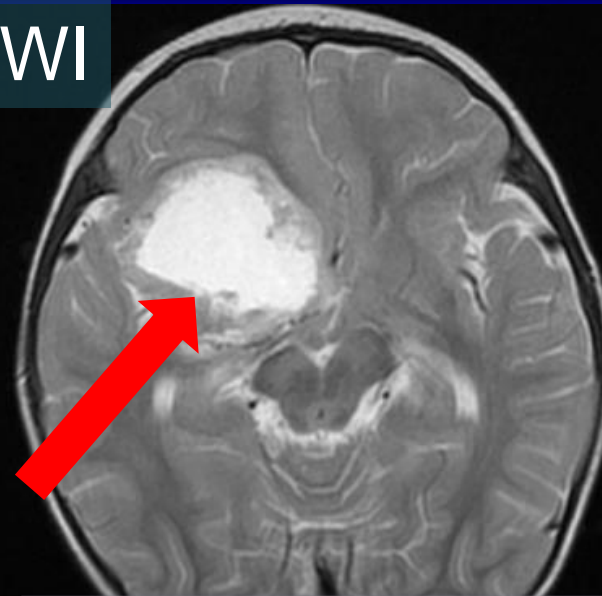


Unenhanced



Calcification

T2WI

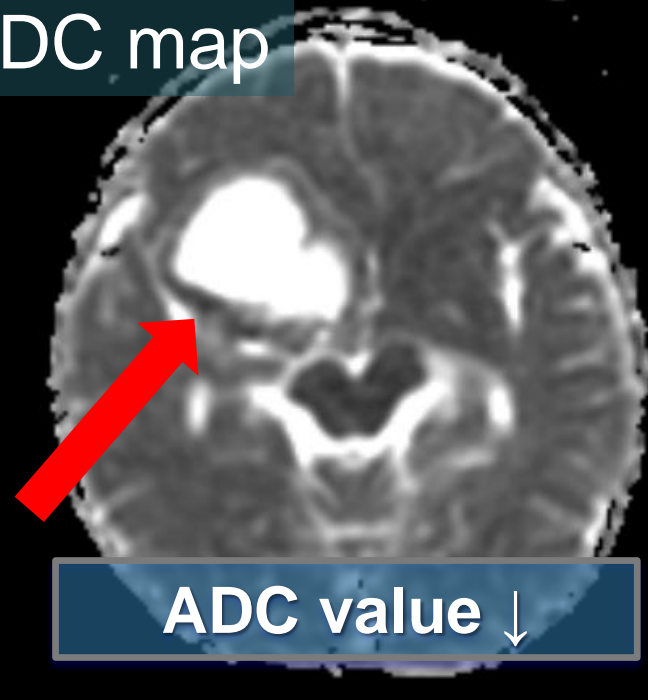


Cystic change

DWI

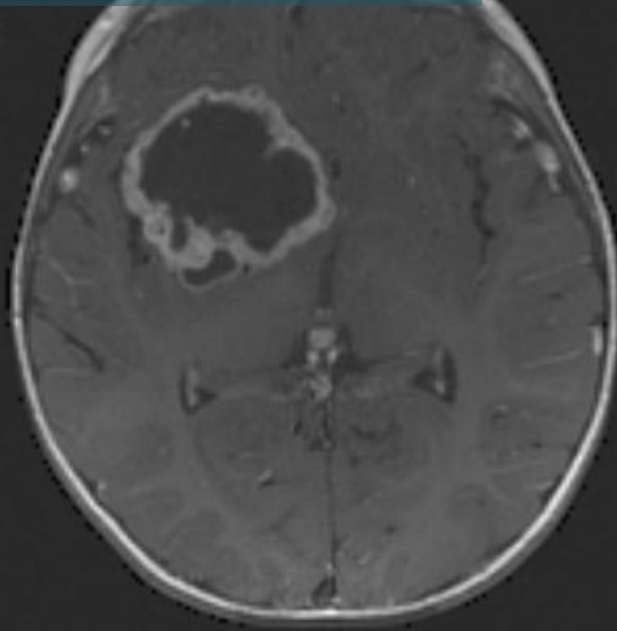


ADC map

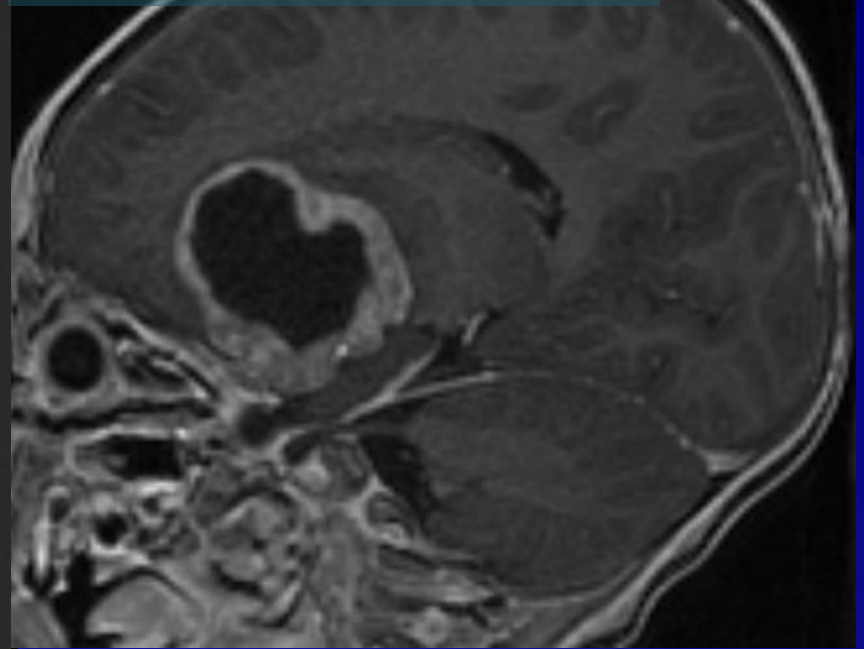


ADC value ↓

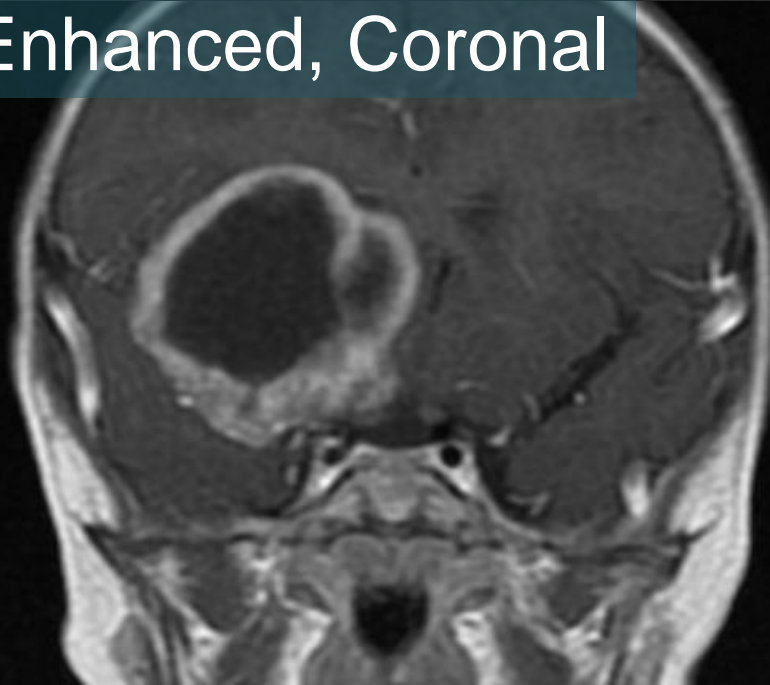
Enhanced T1WI



Enhanced, Sagittal



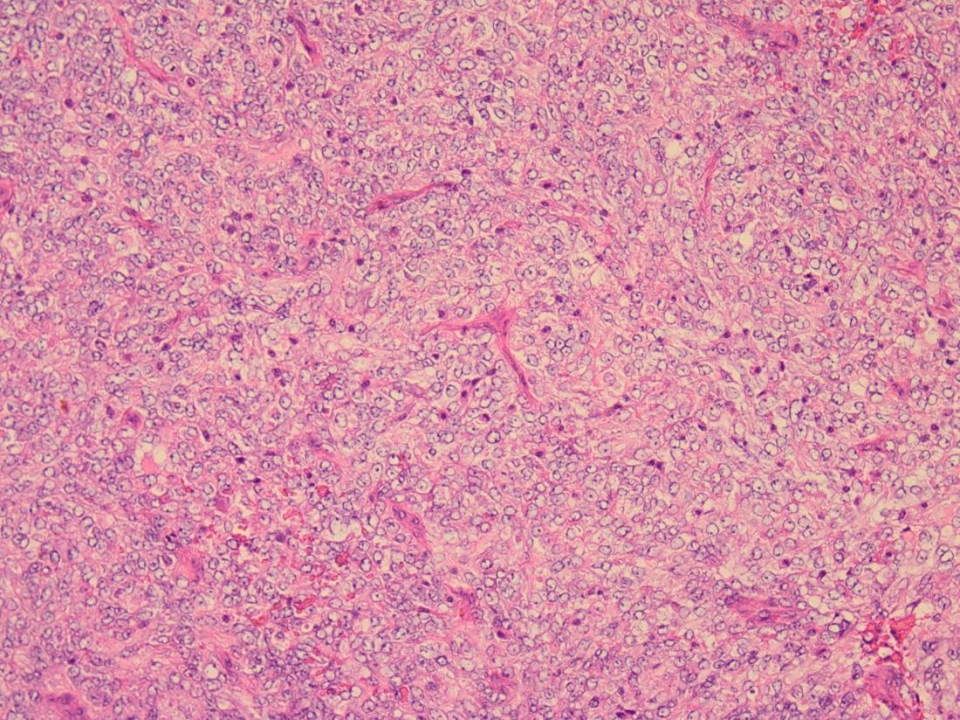
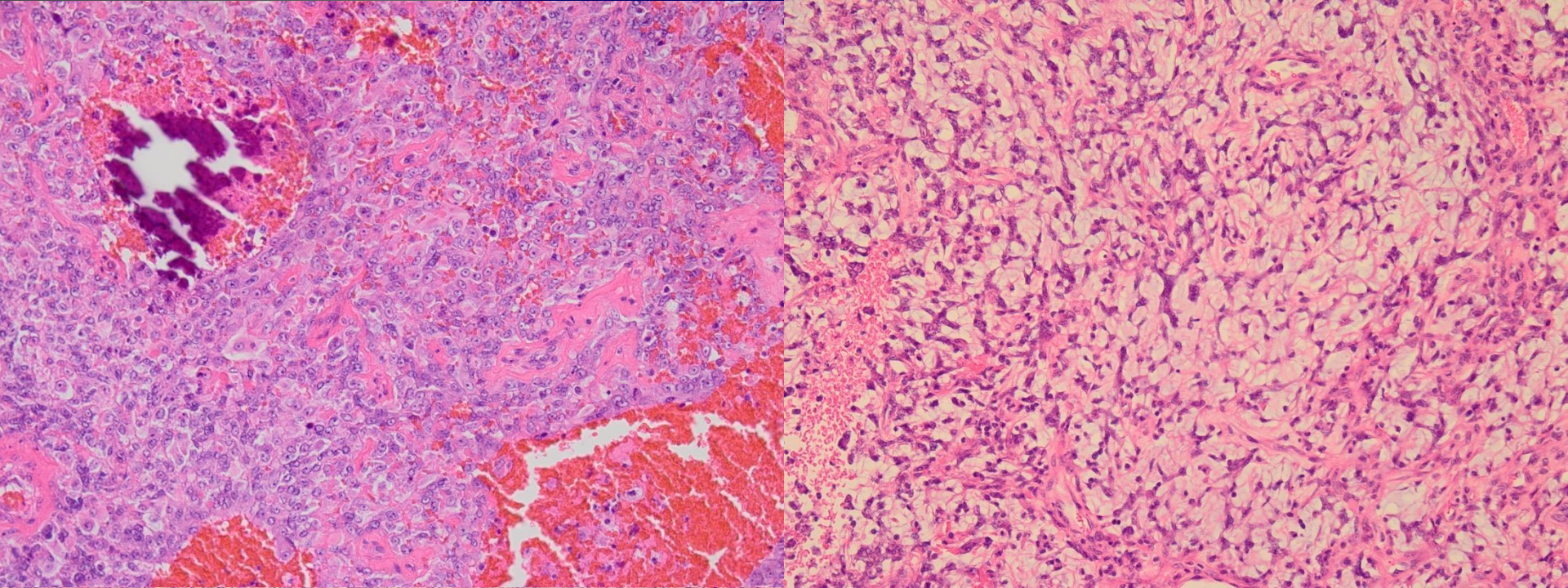
Enhanced, Coronal



**Irregular thick wall  
shows contrast-  
enhancement**

# Clinical course

- ✓ She underwent tumor resection, since the tumor seems to cause the symptoms.

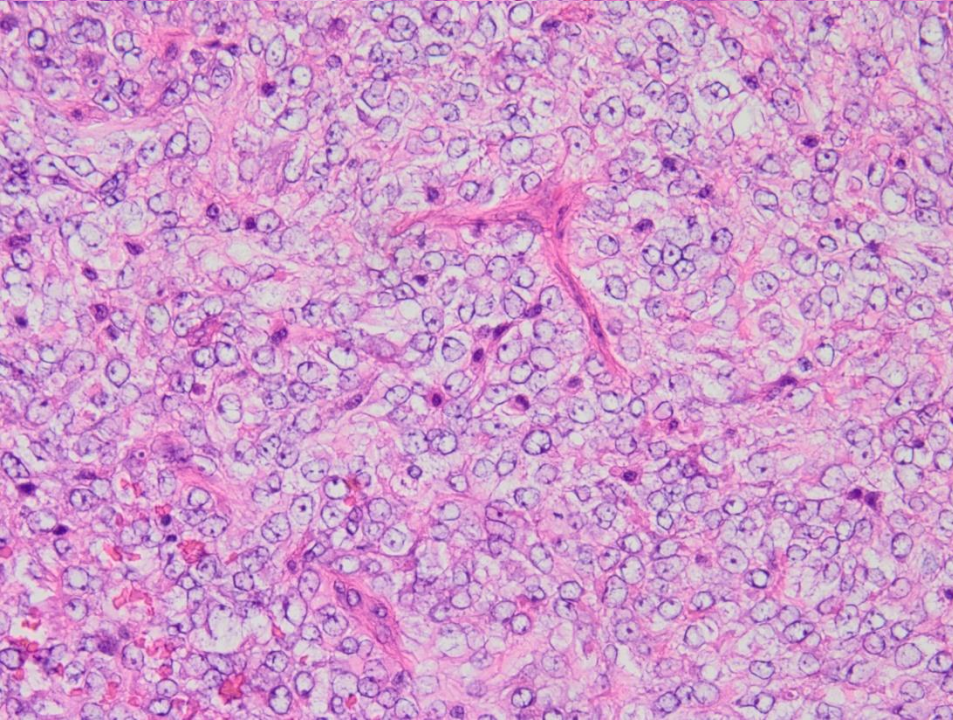
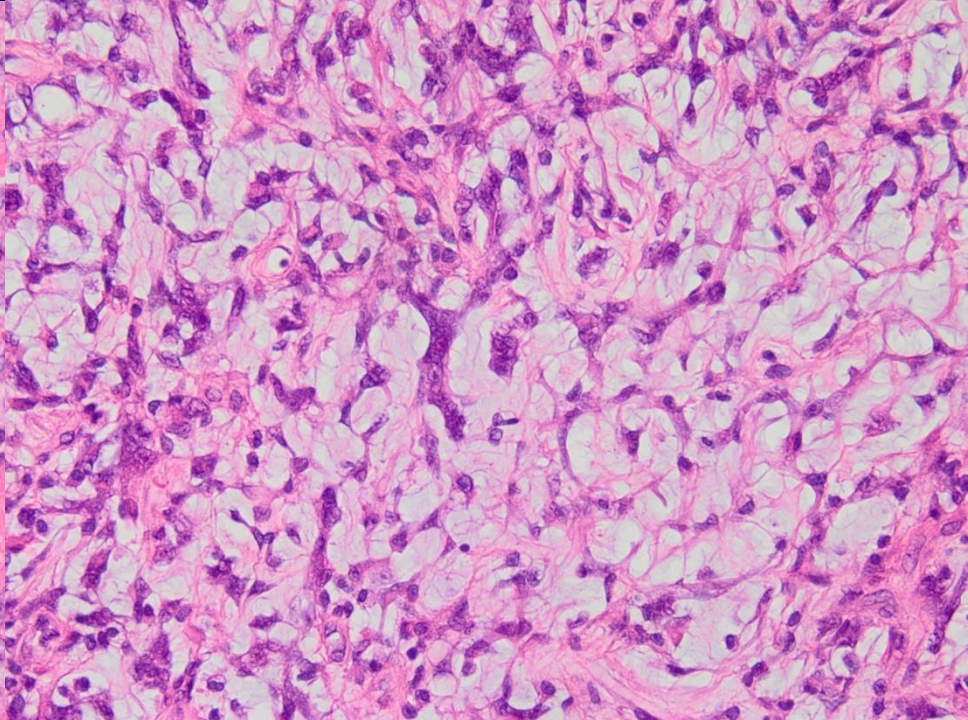
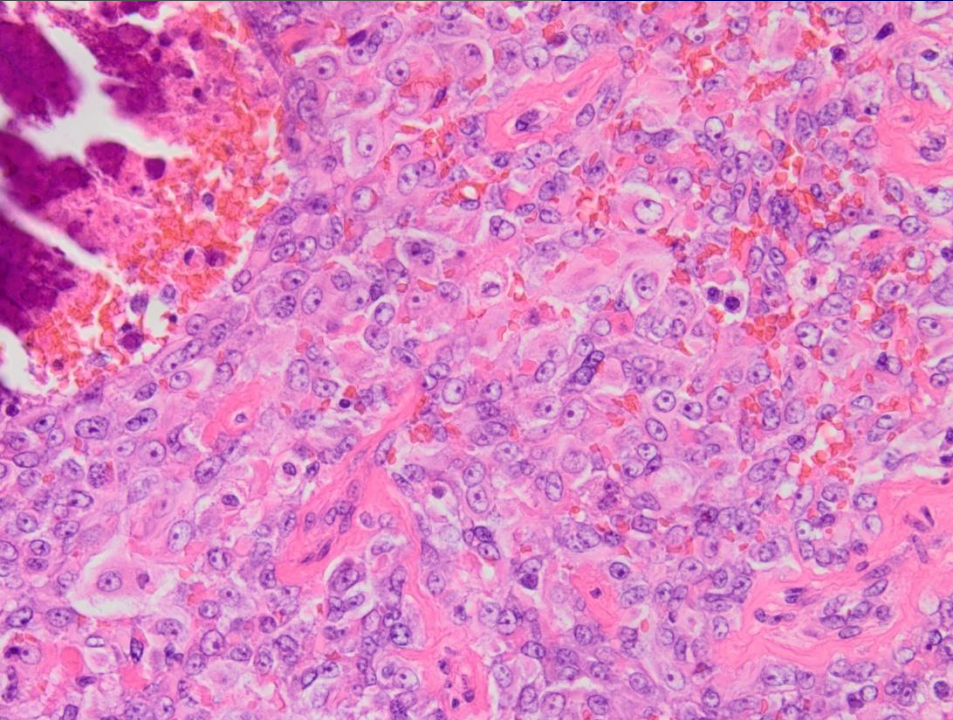


# Low-power field

Rhabdoid

PNET-like

Chordoid  
change

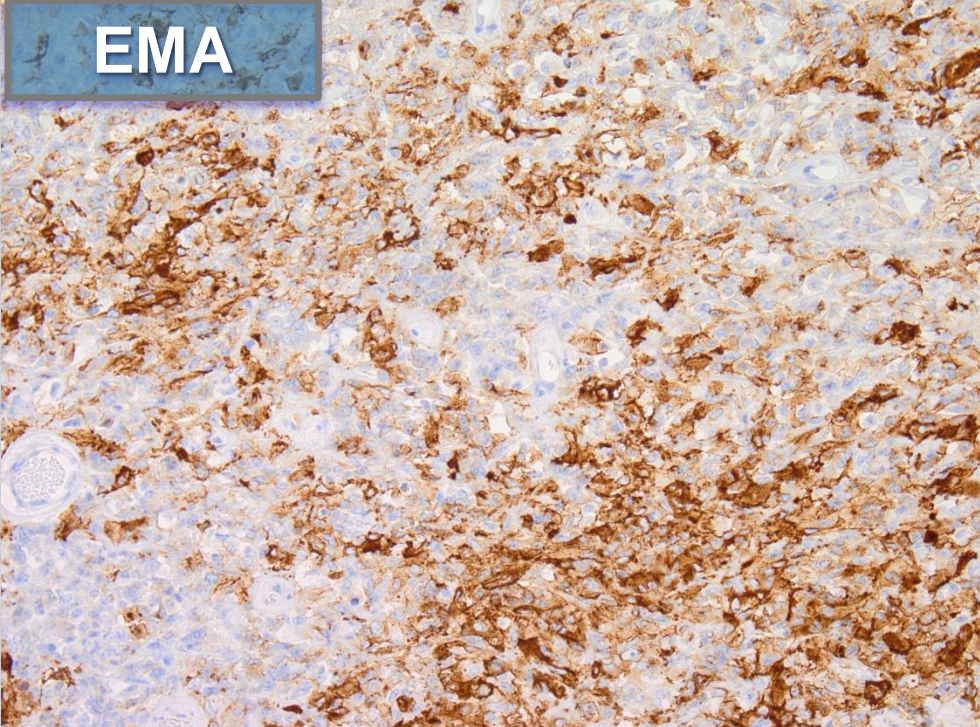
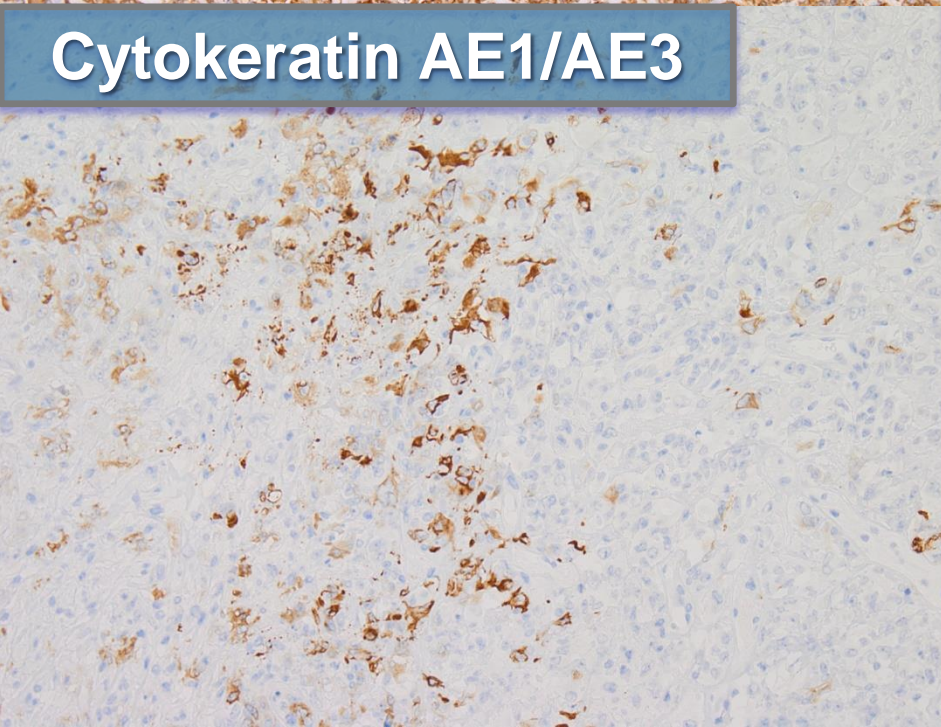
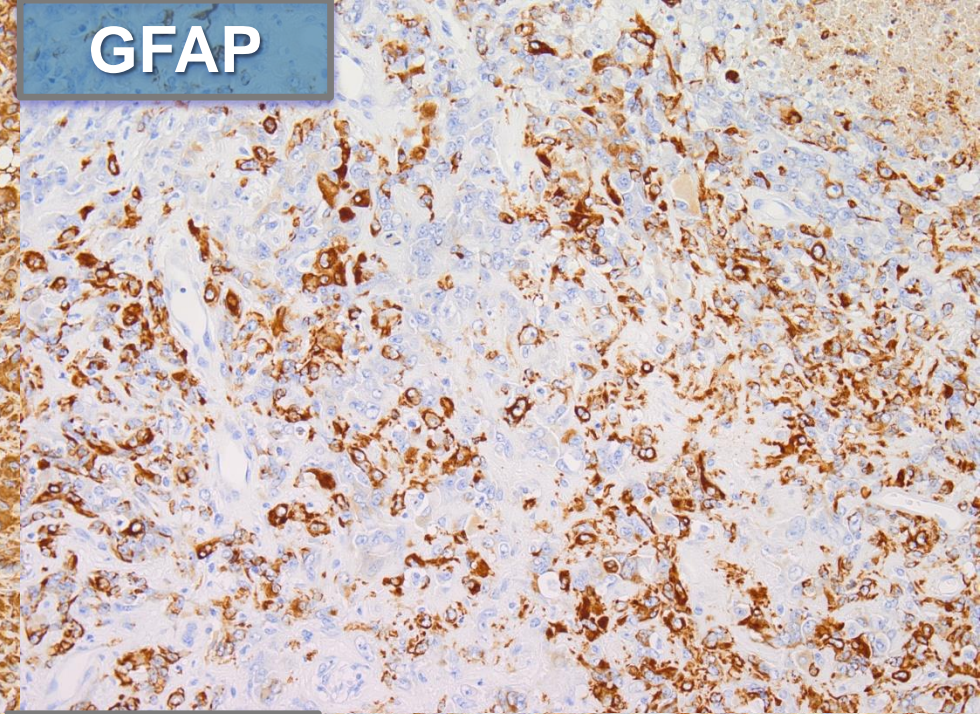
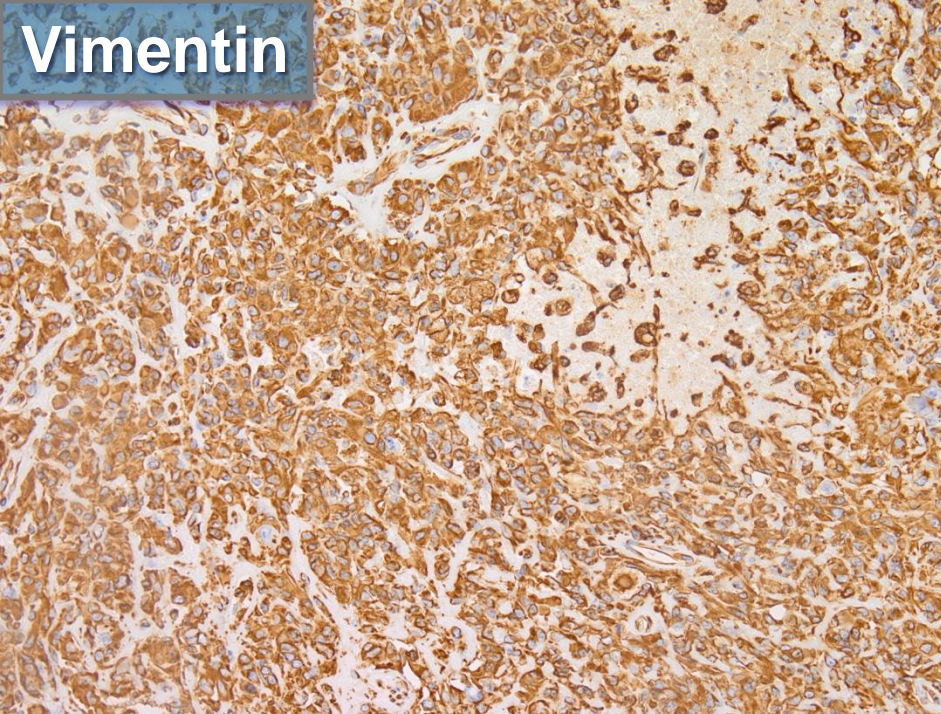


# High-power field

Rhabdoid

PNET-like

Chordoid  
change



30%

40%

60%

Ki-67 labeling index

✓INI-1 (—)

✓Desmin (—)

# Diagnosis

Atypical teratoid / Rhabdoid tumor  
(AT/RT)

G-banding test of the resected tumor  
proved the deletion of chromosome 22.

# Clinical course

- ✓ Patient underwent chemoradiation therapy for 6 months.
- ✓ A renal mass was detected with MRI, suspected malignant rhabdoid tumor of the kidney.
- ✓ Patient passed away 1 year after the diagnosis.

# AT/RT

- ✓ AT/RT contains rhabdoid- as well as varying proportions of PNET-, mesenchymal spindle-shaped-, and/or epithelial-type tumor cells.
- ✓ 1.3% of pediatric primary CNS tumors. (6.7% in the children less than 2 y.o.)
- ✓ Infratentorial (38–65%), supratentorial (27–62%), multiple (4–8%)
- ✓ AT/RT has overlapping features with PNET. Specific immunohistochemical markers are useful to distinguish from PNET and other tumors.

Meyers S.P., et al. *AJNR* 2006 : 27 : 962-71

Biao J, Xiao Yuan Feng, *Pediatric Radiology* 2013 :43 :1001-1008

# Immunohistochemical fetures

Antibody	%Positive
<b>INI-1</b>	<b>&lt;2%</b>
Epithelial membrane antigen	100
Vimentin	100
Actin	83
Cytokeratin	75
Desmin	23
Glial fibrillary acidic protein	87
Neurofilament protein	50

Meyers S.P., et al. *AJNR* 2006 : 27 : 962-71  
Christelle D, et al, *Cancer* 2012 :118:3812-21

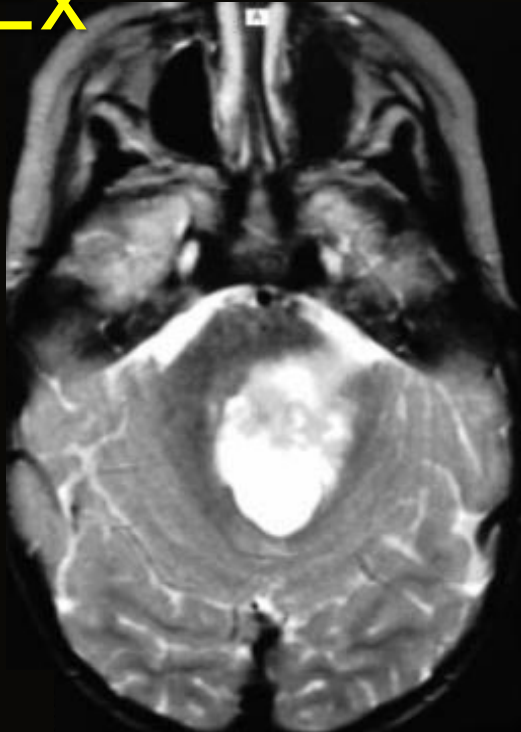
# AT/RT -Radiologic findings-

## ✓ CT findings

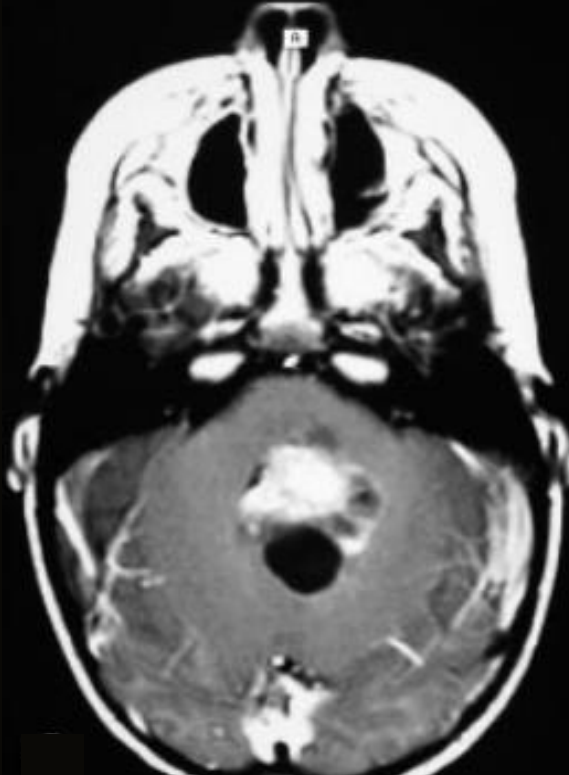
1. Hyperattenuating mass
2. Commonly contains cysts or hemorrhage
3. May contain  $\text{Ca}^{2+}$
4. Obstructive hydrocephalus common
5. Heterogeneous.

# AT/RT -Radiologic findings-

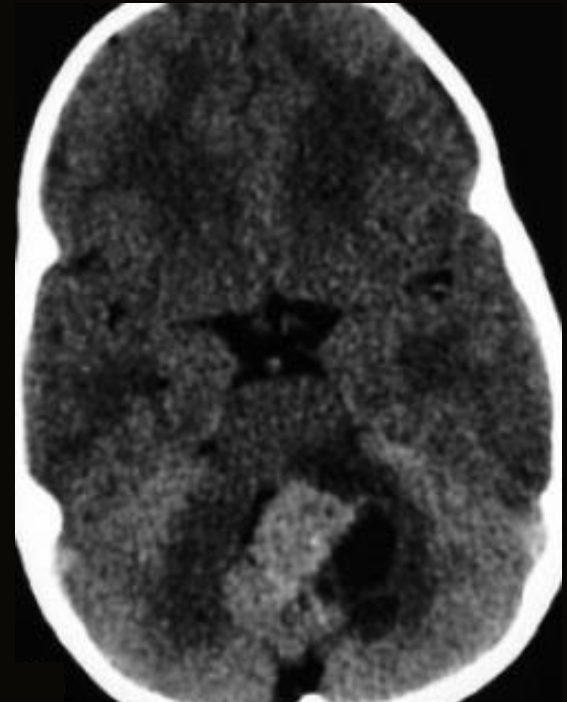
Ex



**T2WI(TR/TE, 5700/90)**  
Cystic change  
Necrosis



**CE-T1WI**  
(TR/TE, 630/22)  
Solid Enhancement  
Cyst – not enhanced



**Tiny calcification**  
Cyst – low attenuation  
Solid – high attenuation

# AT/RT -Radiologic findings-

Tumor	AT/RT	PNET, PNET/MB
Age	<3	<6
Location	Off-mid-line	Mid-line
Hemorrhage	Frequent	Seldom
Cyst Necrosis	Peripheral	Solid mass
Calcification	50%	
DWI	Inhomogeneous	Homogeneous
Enhancement	Heterogeneous+	
MRS	Cho↑, NAA↓, Lip↑	PNET:Lip↑, Lac↑ PNET-MB: Cho↑, Taurine↑

Meyers S.P., et al. *AJNR* 2006 : 27 : 962-71

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# AT/RT -Prognosis-

- ✓ Survival rates: **6–15 months**
- ✓ The presence of disseminated leptomeningeal tumor at diagnosis / staging (**21%–34%**) has been associated with a poor prognosis.
- ✓ Gross total resection improves median survival rates.
- ✓ Gross total or near-complete resection can be performed in fewer than **half of patients**.

Meyers S.P., et al. *AJNR* 2006 : 27 : 962-71

Biao J, Xiao Yuan Feng, *Pediatric Radiology* 2013 :43 :1001-1008

Thank you for your attention